

GUIDELINES FOR THE DIAGNOSIS AND MANAGEMENT OF RECURRENT FSGS IN PEDIATRIC TRANSPLANT RECIPIENTS

I. PURPOSE

To outline guidelines for the diagnosis and management of recurrent FSGS following renal transplantation in children.

II. SETTING

In-patient and out-patient

III. DEFINITIONS

Recurrent FSGS is diagnosed if any of the following are seen post-tx:

- Nephrotic range proteinuria: >3.5 g/24 hr or UPC >3 g/g in adults and FMV or 24 hr urine protein/Cr ratio of > 2g/g or >3+ on dipstick in children AND hypoalbuminemia of < 3 g/dL
- Allograft biopsy shows FSGS pattern or injury and widespread effacement of podocytes
- Early recurrence occurs within 48 hr; intermediate in 2-30 days, late recurrence may be after 3 months post-tx.
- Risk factors include progression to ESRD within 3 yr after dx of FSGS or rFSGS in prior tx

IV. GUIDELINES

Evaluation:

- Monitor urine protein and serum Cr daily for 1 week; twice a week in week 2, weekly for 4 weeks, monthly for the first year and every 3 months thereafter; preferably using the FMV sample.

Treatment of rFSGS:

- Prophylactic TPE or rituximab pre-tx is NOT recommended
- Prompt initial therapy with intensive TPE should be started - daily for 3 days then three times a week for 2 weeks. TPE can be stopped after reduction of proteinuria to < 1 g/d or UPC <1.
- Consider monitoring IgG levels and IVIG if infectious complications
- Alternatively Immunoabsorption may be used; 2.5-3 plasma volumes daily x 1 week followed by QOD for 2 weeks, then BIW for 2 more weeks.
- TPE or IA should also be used for relapse after stopping above Rx.
- CNI may be changed to CsA with high target levels (2 hr peak range 1200-1400 ng/ml until remission)
- Steroids may be used to enhance effect
- Rituximab should be considered; TPE should be held for 48 hr post rituximab dose to prevent drug removal.
- Consider conversion of MMF to oral Cytoxan for 3 months
- LDL-A may be used in pts refractory to TPE or IA

IV. PROCEDURE

Write procedure here (an ideal series of steps that should be followed in a regular, definite order to fulfill policy)

VI. REFERENCES

Post-transplant recurrence of focal segmental glomerular sclerosis: consensus statements; Rupesh Raina, Swathi Jothi, Dieter Haffner, Michael Somers, Guido Filler, Prabhav Vasistha, Ronith Chakraborty, Ron Shapiro, Parmjeet S. Randhawa, Rulan Parekh et al: Kidney International, Volume 105, Issue 3, March 2024, Pages 450-463

VII. REVIEWED BY

Lead developer: Arundhati Kale, MD

Lavjay Butani, MD

Stephanie Nguyen, MD

Maha Haddad, MD

Gia Oh, MD

Machi Kaneko, MD

VIII. REVIEWED DATE and REVIEW CYCLE: June 21, 2024

Medical Legal Disclaimer:

Welcome to the UC Davis Health, Department of Pediatrics, Clinical Practice Guidelines Website. All health and health-related information contained within the Site is intended chiefly for use as a resource by the Department's clinical staff and trainees in the course and scope of their approved functions/activities (although it may be accessible by others via the internet). This Site is not intended to be used as a substitute for the exercise of independent professional judgment. These clinical pathways are intended to be a guide for practitioners and may need to be adapted for each specific patient based on the practitioner's professional judgment, consideration of any unique circumstances, the needs of each patient and their family, and/or the availability of various resources at the health care institution where the patient is located. Efforts are made to ensure that the material within this Site is accurate and timely but is provided without warranty for quality or accuracy. The Regents of the University of California; University of California, Davis; University of California, Davis, Health nor any other contributing author is responsible for any errors or omissions in any information provided or the results obtained from the use of such information. Some pages within this Site, for the convenience of users, are linked to or may refer to websites not managed by UC Davis Health. UC Davis Health does not control or take responsibility for the content of these websites, and the views and opinions of the documents in this Site do not imply endorsement or credibility of the service, information or product offered through the linked sites by UC Davis Health. UC Davis Health provides limited personal permission to use the Site. This Site is limited in that you may not:

- Use, download or print material from this site for commercial use such as selling, creating course packets, or posting information on another website.
- Change or delete propriety notices from material downloaded or printed from it. · Post or transmit any unlawful, threatening, libelous, defamatory, obscene, scandalous, inflammatory, pornographic, or profane material, any propriety information belonging to others or any material that could be deemed as or encourage criminal activity, give rise to civil liability, or otherwise violate the law.
- Use the Site in a manner contrary to any applicable law.

You should assume that everything you see or read on this Site is copyrighted by University of California or others unless otherwise noted. You may download information from this Site as long as it is not used for commercial purposes, and you retain the proprietary notices. You may not use, modify, make multiple copies, or distribute or transmit the contents of this Site for public or commercial purposes without the express consent of UC Davis Health.